Contents

Introduction

Idiopathic pulmonary fibrosis (IPF) is a chronic, fibrosing interstitial pneumonia of unknown cause that is associated with radiological and histologic features of usual interstitial pneumonia (UIP). It occurs primarily in older adults, is characterized by progressive worsening of dyspnea and lung function, and has a poor prognosis. Diagnosis and management of IPF were addressed in prior guidelines (1–3). A formal American Thoracic Society (ATS) and European Respiratory Society (ERS) proposal process determined that several topics from the previous guidelines warrant reassessment, including the following: radiological and histopathological features of UIP, diagnostic criteria, diagnostic and treatment approaches, and prior evidence-based recommendations about antacid medications and transbronchial lung cryobiopsy (TBLC). In addition, it was decided that new questions about antireflux surgery and genomic classifier testing should be addressed.

The acceptance of antifibrotic therapies for IPF led to the investigation of such therapies in other fibrotic lung diseases. While the IPF guidelines were being updated, a clinical trial reporting a beneficial effect of antifibrotic medication in interstitial lung diseases (ILDs) other than IPF that manifest progressive pulmonary fibrosis (PPF) was published (4, 5), prompting a paradigm shift toward an en bloc approach to antifibrotic therapy. Given the importance and timeliness of the issue, the guideline committee was approved to expand its scope to also define progression of pulmonary fibrosis and to decide whether the en bloc approach to antifibrotic therapy should continue, or whether therapy should be restricted to specific types of progressive ILD.

These guidelines for the diagnosis and treatment of IPF and other types of PPF are the result of a collaboration among the ATS, ERS, Japanese Respiratory Society (JRS), and Asociación Latinoamericana de Tórax (ALAT). They are intended to provide the basis for rational, informed decisions. The recommendations should never be considered absolute requirements by anyone who evaluates the actions of a healthcare professional.

Methods

Methods including conflict-of-interest management were established *a priori* and are described in the online supplement. The document can be conceptualized in two parts. Narrative portions (e.g., radiological criteria, histopathological criteria, physiological criteria, definitions) were created using consensus by discussion. Guideline portions address specific questions related to TBLC, genomic classifier testing, antacid medication, antireflux surgery for IPF, and pirfenidone and nintedanib for PPF. These sections are compliant with the Institute of Medicine standards for trustworthy guidelines (6) and yield recommendations that were informed by systematic reviews and were formulated and graded using the Grading of Recommendations, Assessment, Development and Evaluation (GRADE) approach (7) (Table 1).

Evidence-based recommendations were formulated by discussion followed by voting. Briefly, committee members were provided the following options: a strong recommendation for a course of action, a

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